Unusual Right Ventricular Thrombus in a Woman with Hughes-Stovin Syndrome

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A 28-year-old woman with no prior pathologic antecedent was admitted to the internal medicine department for a 2-month history of intermittent fever and swollen lower limbs. Physical examination showed bilateral pitting leg edema with no inflammatory skin changes. Her body temperature was above 38°C. Vascular echo-Doppler revealed a bilateral primitive iliac vein thrombosis extending to the vena cava. Echocardiography objectified a right ventricular rounded hyperechogen mass, suggesting myxoma or thrombus. Thoracic angio-CT-scan showed an aneurysm of the inferior branch of right pulmonary artery, surrounded by parenchymal ground-glass opacity in favour of vasculitis (Picture 1), and confirmed the diagnosis of right ventricular thrombus (Picture 2). Biologically, CRP was at 9 mg/L with no other laboratory assay abnormality. Thrombophilia screening and immunological tests were negative. HLA-B51 was positive. In the absence of suggestive manifestations (skin or ocular lesions) of Behcet’s disease (BD), the diagnosis of Hughes-Stovin syndrome (HSS) was made. The patient was treated with bolus cyclophosphamide with heparin relayed by antivitamin K, under medical supervision.

HSS is a rare entity characterized by the association of pulmonary artery aneurysms and peripheral venous thrombosis, which was first described in 1959 and classically affects men at a young age (1). The aetiology of HSS is still unknown; however it is thought to be a clinical variant manifestation of BD in which the typical symptoms such as oral

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Picture 1. Sagittal thoracic angio-CT-Scan: right pulmonary artery aneurysm (arrow), surrounded by parenchymal ground-glass opacity (arrowhead).

Picture 2. Axial thoracic angio-CT-Scan: right ventricular thrombus (arrow).
or genital ulceration, are mostly absent. Cardiac thrombus, mainly in the right ventricle, was exceptionally reported in HSS (1, 2).

References
